Rosai-Dorfman Disease Presenting as Isolated Cervical Lymphadenopathy in a 12-Year-Old Female: A Case Report

Nayan Sapkota,[1] Sameer Karmacharya,[1] Bikash Lal Shrestha,[1] Neha Amatya[1]

[1]Department of ENT-HNS, Dhulikhel Hospital, Kathmandu University Hospital, Dhulikhel, 45210, Kavre, Nepal

✉ **Corresponding Author:**

Dr Nayan Sapkota

Email ID: nayansapkota105@gmail.com

Abstract

Background

Rosai-Dorfman disease (RDD) is a rare, benign disorder characterized by painless, massive cervical lymphadenopathy. First described in 1965, it commonly presents with fever, leukocytosis, elevated ESR, and hypergammaglobulinemia. Diagnosis is confirmed by histopathology showing histiocyte proliferation with emperipolesis and positive S-100 and CD68 staining.

Case Presentation

A 12-year-old girl presented with a gradually enlarging left cervical lymph node. Fine-needle aspiration cytology and excisional biopsy confirmed RDD. After surgical excision, she was treated with steroids, leading to regression of lymphadenopathy.

Conclusions

RDD generally has a benign, self-limiting course. However, careful monitoring is essential due to possible relapse or extranodal involvement. Early diagnosis and appropriate management yield favorable outcomes.

Keywords: *Case Report, Histiocytosis, Juvenile, Lymph Nodes, Rosai-Dorfman Disease, Sinus Histiocytosis.*

Introduction

Rosai-Dorfman disease (RDD), or sinus histiocytosis with massive lymphadenopathy (SHML), is a benign, self-limiting disorder marked by painless cervical lymph node enlargement[1]. Classic symptoms include fever, leukocytosis, and hypergammaglobulinemia[2]. Notably, 40% of cases exhibit extranodal involvement in organs such as skin, eyes, nervous system, bone, gastrointestinal tract, and more, often mimicking other conditions[2-6]. Diagnosis of nodal RDD is typically straightforward on H&E staining with supportive immunohistochemistry, but extranodal presentations or those with fibrosis and minimal emperipolesis can be challenging[7]. Our case is notable for presenting as isolated unilateral cervical lymphadenopathy in a 12-year-old without systemic symptoms or extranodal involvement. FNAC followed by excisional biopsy confirmed RDD, highlighting the importance of histological evaluation even in clinically ambiguous cases. The rarity of such isolated pediatric presentations without systemic signs makes this case

unique and worthy of reporting.

Case Report

12-year-old female presented to the Otorhinolaryngology outpatient department of Dhulikhel Hospital with a one-year history of swelling over the left upper lateral aspect of the neck (Figure 1). The swelling was painless and gradually progressive, with no associated history of fever or cough.

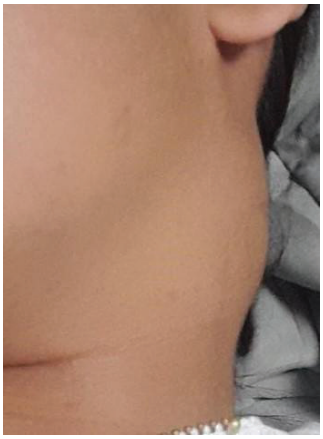

Figure 1. ***Rosai-Dorfman disease: A 12-year-old girl with left cervical lymphadenopathy***

Clinical examination revealed multiple enlarged left cervical lymph nodes, ranging in size from 1×2 cm to 2×2 cm. The overlying skin appeared normal, with no change in color or local rise in temperature. The lymph nodes were non-tender, firm to hard in consistency, mobile, with smooth and regular surfaces, and not adherent to the skin. No other lymphadenopathy was detected.

Ultrasonography of the neck showed multiple enlarged lymph nodes with loss of central fatty hilum at levels IB and V on the left side, the largest measuring 25×11 mm. CT scan of the neck revealed multiple homogeneously enhancing, discrete enlarged lymph nodes in the left cervical region (levels IB and V), the largest measuring 25×22 mm.

Fine-needle aspiration cytology (FNAC) of the left cervical lymph nodes showed numerous histiocytes with eccentrically placed round-to-oval nuclei, prominent nucleoli, and abundant pale cytoplasm, with evidence of lymphophagocytosis (emperipolesis). The cytological diagnosis was Rosai-Dorfman disease (RDD). The patient was initially managed with anti-inflammatory medication and kept under regular follow-up. However, after two months, the swelling did not regress, and an excisional biopsy was planned. Under general anaesthesia, a single soft, yellowish, well-circumscribed encapsulated globular lymph node mass measuring 3×3 cm was excised (Figure 2).

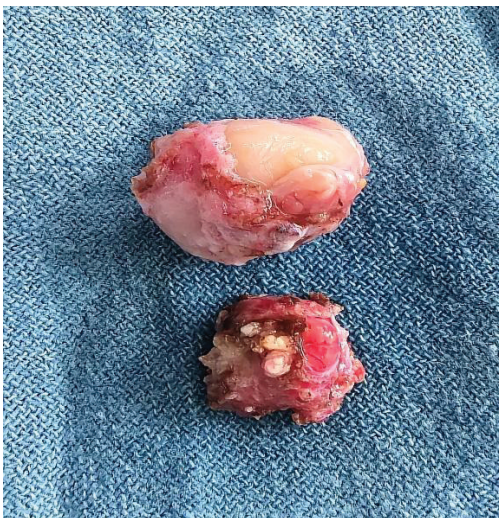

Figure 2. *Photograph of excised lymph node*

Histological examination revealed encapsulated lymphoid tissue with effaced architecture (Figure 3). The distended sinuses contained a prominent population of histiocytes characterized by large round-to-oval nuclei, prominent nucleoli, and abundant cytoplasm showing emperipolesis, engulfment of intact lymphocytes.

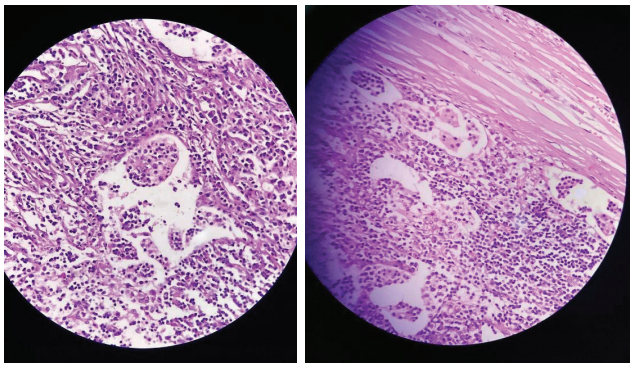

Figure 3. *Photomicrograph showing lymphoid tissue with effaced architecture and emperipolesis, suggestive of sinus histiocytosis*

The intervening stroma showed an increased number of plasma cells, blood vessels, and areas of haemorrhage. The final diagnosis was sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease). Postoperatively, the patient was treated with oral corticosteroids for two weeks, after which progressive regression of the lymph node swelling was observed (Figure 4).

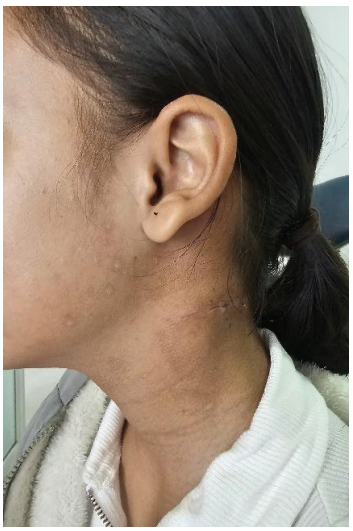

Figure 4. *Post-operative photograph after excision of the lymph nodes*

The patient remains under regular follow-up, with no evidence of recurrence to date.

Conclusion

Rosai-Dorfman disease (RDD) is a rare, benign histiocytic disorder that can mimic more serious pathologies, particularly when presenting as isolated lymphadenopathy without systemic symptoms. Early recognition based on characteristic cytological features especially emperipolesis, supported by histopathology and immunohistochemistry, is essential for accurate diagnosis and appropriate management. Our case highlights an uncommon presentation of RDD as unilateral cervical lymphadenopathy in a pediatric patient without systemic or extranodal involvement, which makes it clinically unique and diagnostically challenging. Surgical excision combined with a short course of corticosteroids led to complete resolution. This case reinforces the importance of considering RDD in the differential diagnosis of persistent cervical lymphadenopathy in children and the value of a multidisciplinary approach in ensuring effective management and follow-up.

Consent

Written informed consent was obtained ensuring patient's anonymity.

Declaration of competing interest

There are no conflicts of interest.

Acknowledgement

N/A

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